Int J Clin Exp Med 2018;11(9):9152-9159
www.ijcem.com /ISSN:1940-5901/IJCEM0068042

Original Article

Diagnoses and treatment decision in anomalous systemic arterial supply to basal segments without sequestration

Yunfeng Shi[1*], Benquan Wu[1*], Jie Qin[2], Hui Liu[1], Jian Zhang[3], Tiantuo Zhang[1]

[1]Department of Internal Medicine, Division of Respiratory and Critical Care, The Third Affiliated Hospital of Sun Yat-Sen University, Guangzhou, P.R. China; Departments of [2]Radiology, [3]Cardiothoracic Surgery, The Third Affiliated Hospital of Sun Yat-Sen University, Guangzhou, P.R. China. [*]Equal contributors.

Received October 27, 2017; Accepted May 24, 2018; Epub September 15, 2018; Published September 30, 2018

Abstract: *Objective:* To conclude the diagnoses and treatment decision in anomalous systemic arterial supply to basal segments without sequestration. *Methods:* Clinical manifestations, CT scanning, diagnoses and treatment of 6 cases from 2010 to 2013 were reported together with literature review. *Results:* All 6 cases were diagnosed by high resolution CT, and were confirmed in operation. It presented clinical features as male gender and hemoptysis. CT showed one anomalous systemic artery arising from descending thoracic aorta supplying the left lower lobe. 1 case who was comitant with von Willebrand disease survived after endovascular embolization, 4 of 5 cases were cured by surgical treatment and 1 died of postoperative bleeding. *Conclusions:* Definite diagnoses can be made by means of high resolution CT. For patients with complicated condition, endovascular embolization could be a safer alternative.

Keywords: Anomalous systemic arterial supply, high resolution computed tomography, video-assisted thoracoscopic surgery, endovascular embolization

Introduction

Anomalous systemic arterial supply to basal segments without bronchial pulmonary sequestration is a rare congenital anomaly. However, it has arisen more attention as the use of high-resolution CT (HRCT) in these years. The treatment modality is still controversial regarding whether endovascular emboliztion is the first choice for all cases. We reported 6 patients who were diagnosed by HRCT and treated by operation, then literature was reviewed.

Case 1

A 31-year-old male was admitted to our hospital because of repeated hemoptysis for 7 months. About 5 mL bright red blood was coughed out each time when hemoptysis occured. There was no fever, night sweat, palpitation, chest pain, pus sputum and breathlessness. He used to suffer gingival bleeding, subcutaneous hemorrhage and epistaxis since childhood, even had upper gastrointestinal hemorrhage once. He was non-smoker, denied positive family history. His life signs were stable. There was no positive sign in physical examination.

In blood studies, no inflammatory findings were noted. Platelet test was also normal. However, activity of coagulation factors decreased. Hemophilia screening tests showed that coagulation factor VIII activity was 28% (reference range from 60% to 150%), factor IX activity was 62% (reference range from 60% to 150%), VWF activity was 18% (reference range from 50% to 160%), factor XI activity was 74% (reference range from 60% to 140%), and factor XII activity was 46% (reference range from 60% to 140%). During bronchoscopy examination, the patient continued expectorating dark blood from bronchi in left lower lobe, but his bronchial tree was normal without sequestration. Contrast-enhanced chest CT and volume reconstruction (VR) were performed, the result showed a dilated anomalous artery (15 mm in diameter) arising from lateral aspect of descending aorta supply to left basal segments (**Figure 1**).

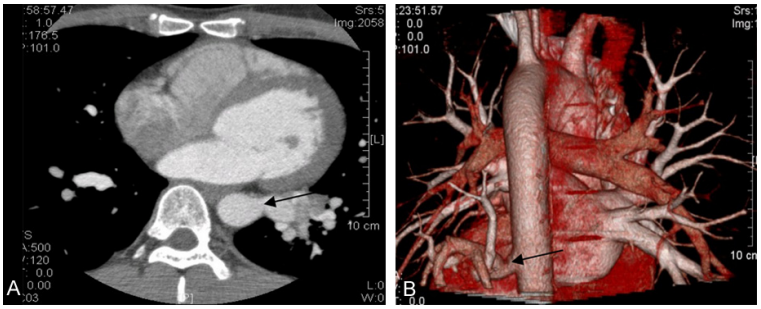

Figure 1. Contrast-enhanced CT scan at mediastinal window setting at level of left atrium showed dilated anomalous artery (15 mm in diameter) arising from lateral aspect of descending aorta (A). Volume reconstruction (VR) (B) showed dilated anomalous artery arising from lateral aspect of descending aorta and a small left inferior pulmonary artery.

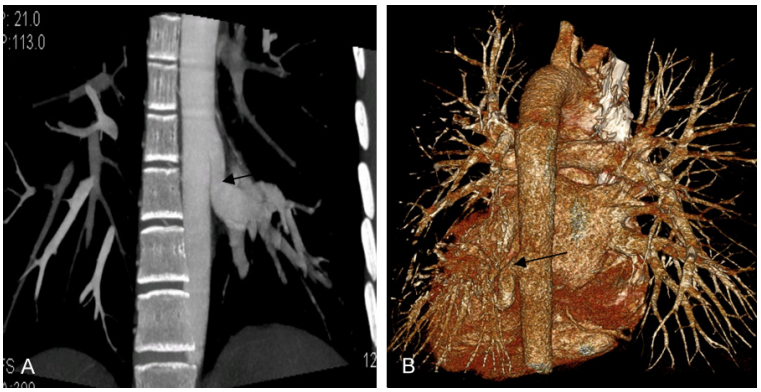

Figure 2. Maximum intensity projection (MIP) (A) and VR (B) showed dilated anomalous artery (10 mm in diameter) arising from lateral aspect of descending aorta. The interlobar artery distal to the origin of the superior segmental artery was absent. Venous return was via the large inferior pulmonary vein into the left atrium. No direct communications existed between the anomalous systemic artery and the veins of the basal segments.

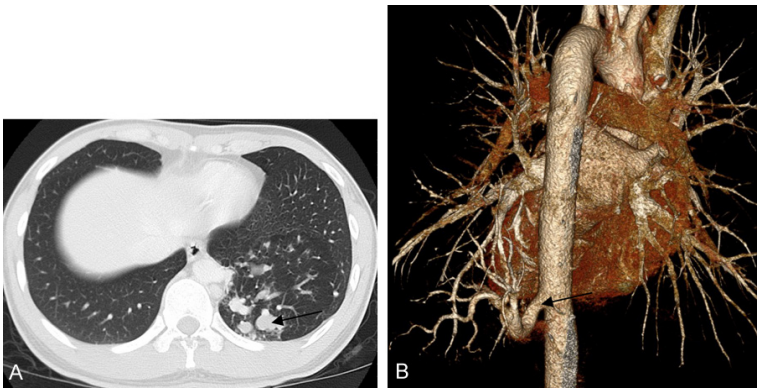

Figure 3. CT scan at lung window setting showed anomalous artery (10 mm in diameter) and its branches paralleling normal bronchi in small left lower lobe (A). VR (B) showed dilated anomalous artery arising from lateral aspect of descending aorta. No direct communications existed between the anomalous systemic artery and the veins of the basal segments.

The patient's final diagnoses was anomalous systemic arterial supply to left basal segments comitant with von Willebrand disease.

The patient was prescribed with fresh frozen plasma and cryoprecipitate to stop hemoptysis. He refused any surgical or endovascular therapy because of potential complications in the beginning. Then he accepted endovascular embolization with coil in Nanfang Hospital Affiliated to Southern Medical University one year later. The operation was successful. The anomalous systemic artery was completely obstructed. Hemoptysis ceased and no side effect emerged during follow-up for more than 5 years. As far as we knew,this was the first case reported of such a anmomaly comitant with von Willebrand disease and cured by embolization with coil.

Case 2

A 29-year-old male presented with recurrent hemoptysis for 5 years. About 15 mL bright red blood was coughed out each time for three consecutive days. He was non-smoker, and had no special past history. Physical examination and laboratory findings showed no abnormalities. Bronchoscopy revealed no abnormality in bronchial tree.

Chest CT showed that a dilated anomalous artery arising from lateral aspect of descending aorta supplying to left basal segments, the interlobar artery distal to the origin of the superior segmental artery was absent, and venous return was via the large inferior pulmonary

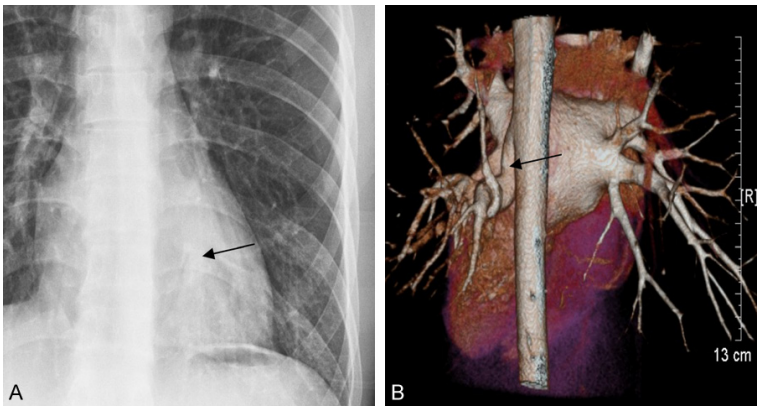

Figure 4. Chest X ray showed a vessel-like opacity in the left lower lobe (A). VR showed a sigmoid-shaped anomalous systemic artery (11 mm in diameter) originating from the lower descending thoracic aorta with three branches. The left inferior pulmonary vein in the medial part of the left lower lobe was noted (B).

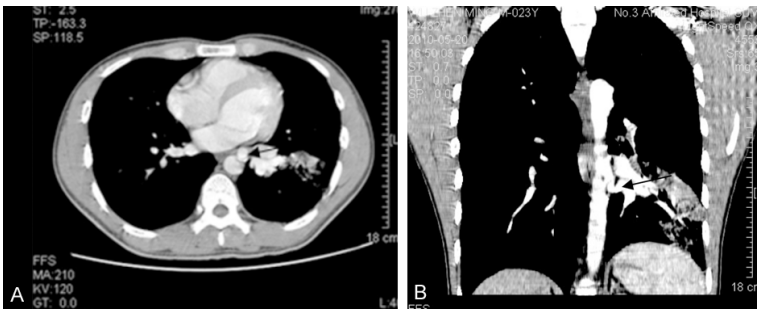

Figure 5. Contrast-enhanced CT scan at mediastinal window setting showed the anomalous systemic arteries (10 mm in diameter) and its branches accompanying the normal basal segmental bronchi and the accompanying interlobar artery is absent (A). MIP showed dilated anomalous artery arising from lateral aspect of descending aorta. Venous return was via the large inferior pulmonary vein into the left atrium. No direct communications existed between the anomalous systemic artery and the veins of the basal segments (B).

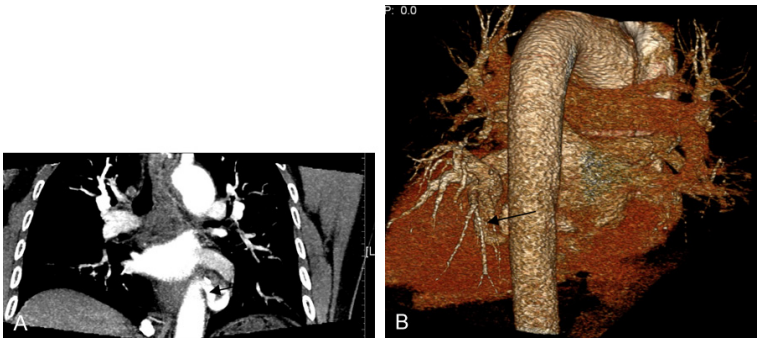

Figure 6. MIP (A) and VR (B) showed dilated anomalous artery (6 mm in diameter) arising from lateral aspect of descending aorta. Venous return is via the large inferior pulmonary vein into the left atrium. No direct communications exist between the anomalous systemic artery and the veins of the basal segments.

vein into the left atrium (**Figure 2**).

The patient underwent left lower lobectomy with ligation of the anomalous artery by Video-assisted Thoracoscopic Surgery (VATS) one year later in the First Affiliated Hospital of Guangzhou Medical University. The operation was uneventful. No hemoptysis recurred during follow-up.

Case 3

A 27-year-old male, non-smoker, was referred to our hospital because of repeated cough and bloody sputum for 8 months. There was no fever, chest pain and tightness. He accepted antibiotics therapy including azithromycin and levofluxacin, but it was in vain. His past history was negative. Physical examination and laboratory findings were normal as well as bronchoscopy examination. Chest CT scanning found anomalous systemic arterial supplying to normal basal segments of the left lower lobe (**Figure 3**).

The patient accepted left lower lobectomy with ligation of the anomalous artery by VATS in the First Affiliated Hospital of Sun Yat-sen University. The operative course was satisfactory and he was asymptomatic in follow-up.

Case 4

An 18-year-old non-smoker male was referred to our hospital because of cough and hemoptysis for 2 months. Hemoptysis ceased after haemostasis and anti-infection therapy in local hospital. He had no tuberculosis and congenital heart disease. Physical ex-

amination and laboratory findings were within normal range at admission. Bronchoscopy showed no bronchial pulmonary sequestration. Chest CT showed a sigmoid-shaped anomalous systemic artery originating from the lower descending thoracic aorta with three branches supplying to basal segments of the left lower lobe (**Figure 4**).

Left lower lobectomy with ligation of the anomalous artery was performed under VATS in our hospital. The patient recovered successfully. Hemoptysis disappeared during follow-up.

Case 5

A 23-year-old male, non-smoker, was presented to our hospital with complain of cough, bloody sputum and high fever for one month. He coughed out about 30-40 mL bright red blood each time for 3 days before hospitalization. The patient had no special past history. He had normal physical examination and laboratory findings at admission. Colour Doppler ultrasound showed an arterial branch from descending aorta. Bronchoscopy examination was normal. Anomalous systemic arterial supply to basal segments in the left lower lobe without sequestration was demomstrated by Chest CT (**Figure 5**).

No surgical contraindication was found and the patient underwent left lower lobectomy with ligation of the anomalous artery by VATS in our hospital. Regretfully, the patient died of complication of postoperative bleeding.

Case 6

A 54-year-old non-smoker female was presented to our hospital because of repeated hemoptysis for 4 years. Hemoptysis could be ceased without treatment each time. Her past history was negative. There was no positive sign in physical examination. Bronchoscopy showed no abnormal findings. Chest X-ray showed shadow in left lower lobe. Chest CT showed anomalous systemic artery supplying to left basal segments of the lower lobe (**Figure 6**).

Left lower lobectomy with ligation of the anomalous artery was performed under VATS in Guangdong General Hospital. The patient's postoperative recovery was uneventful, and she was free from hemoptysis during follow-up.

Results

Case summaries

The clinical features of the 6 patients are summarized in **Table 1**. SPSS 20 software was utilized for the inputing, tabulation, and statistical analysis.

Clinical data

The median age of the 6 patients were 25 years (rage 18-54). Five cases (83.3%) were young male and one (16.7%) was old female. All of them complained hemoptysis and 4 patients (66.7%) complained persistent cough simultaneously. Patient 1 had a comitant disease of von Willebrand disease diagnosed this time. Physical examinations were normal. Cardiac murmurs or abnormal electrocardiogram findings were not detected in any of the patients. Decrease of coagulation activity was found in patient 1, there were no special findings in laboratory test in other patients. Bronchoscopy in all 6 cases showed no bronchial sequestration.

All of 6 patients' final diagnoses were made as anomalous systemic arterial supply to the basal segments without pulmonary sequestration by means of medical imageology, using 320-detector CT with volumn reconstruction in our hospital, and were confirmed by surgery or angiopraphy. In all cases, imageology showed that the left basal segments were supplied by one anomalous artery, which arose from the descending thoracic aorta, without pulmonary arteries to the basal segments. The anomalous artery was 6-15 mm in diameter (with a median of 10 mm). No direct communications between the anomalous systemic artery and the veins of the basal segments were noted, and then the blood drained into the left atrium via the large inferior pulmonary vein.

Left lower lobectomy with ligation of the anomalous artery by VATS was peformed in patients 2-6. Four cases showed a good prognosis. Regretfully, patient 5 died of postoperative bleeding. Patient 1 who had comitant disease of von Willebrand disease underwent endovascular embolization with coil 1 year later and achieved a good result.

According to the data of patients 4 and 5, marked capillary dilatation was seen on the vis-

Table 1. Clinical features of the patients

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Sex	male	male	male	male	male	female
Age (year)	31	29	27	18	23	54
Symptom	hymoptysis	cough hemoptysis	cough hemoptysis	cough hemoptysis	cough hymoptysis fever	hymoptysis
Smoker	non	non	non	non	non	non
Family history	negative	negative	negative	negative	negative	negative
Comitant disease	vWD	none	none	none	none	none
Heart murmur	(-)	(-)	(-)	(-)	(-)	(-)
Laterality	left	left	left	left	left	left
Location	basal segment	basal segment	basal segment	basal segment	basal segment	basal segment
Number of aberrant artery	1	1	1	1	1	1
Diameter	15 mm	10 mm	10 mm	11 mm	10 mm	6 mm
Origin	DTA	DTA	DTA	DTA	DTA	DTA
Pulmonary artery	absence	absence	absence	absence	absence	absence
Drainage vein	LIPV	LIPV	LIPV	LIPV	LIPV	LIPV
Pulmonary artery supply	none	none	none	none	none	none
Abnormal of bronchial tree	(-)	(-)	(-)	(-)	(-)	(-)
Sequestrated lung	(-)	(-)	(-)	(-)	(-)	(-)
Treatment	endovascular embolization	ligation lobectomy by VATS	ligation lobectomy by VATS	ligation lobectomy by VATS	ligation lobectomy by VATS	ligation lobectomy by VATS
Histological pathology	NA	NA	NA	Alveolar hemorrage	Alveolar hemorrage	NA
Result	being well	being well	being well	being well	death	being well

DTA, descending thoracic aorta; LIPV, left inferior pulmonary vein; NA, not acquired; vWD, von Willebrand disease.

ceral pleural membrane of the basal segment during operations, without pleural adhesion. Histopathological examination showed that the anomalous artery was elastic vessel, tissue around the artery was rich in collagen fiber. Pulmonary consolidation caused by edema, partial congestion and bleeding was seen. Hemodiderin-laden-macrophages and lymphocytes were noted in the alveoli and interalveolar tissue without fibrous hypertrophy. Alveolar hemorrhage in histological pathology was in accordance with hemoptysis clinically.

Discussion

Anomalous systemic arterial supply to the basal segments without sequestration was a very rare congenital anomaly. It was regarded as Pryce type I sequestration [1]. However, this anomaly has normal bronchial connections and the involved lung is not sequestrated. More and more data showed that anomalous arterial supply was different congenital anomaly from true sequestration [2]. With the rapid development of HRCT and magnetic resonance imaging (MRI), the number of diagnosed and reported cases is increasing. Consensus of diagnoses and treatment is more important than the category of this anomaly, that's why we reviewed this literature.

WU discovered such anomaly was related to Asian ethnicity [3]. Most cases reported were Japanese, Chinese and Indian. Hemoptysis, cough, bloody sputum and dyspnea are the most frequent clinical symptoms in adults. 100% cases we reported had hemoptysis. While the main manifestations in children are heart murmur, respiratory distress, cardiomegaly, or heart failure [4]. The diagnoses should be suspected when a patient suffered recurrent hemoptysis and cough, as well as in the presence of a continuous murmur, cardiomegaly and persistent shadow on chest radiographs, especially for Asian patients [3]. The most difficult differential diagnoses is pulmonary sequestration. Bronchoscopy examination is essential for confirming the presence or absence of a normal tracheobronchial connection.

Conventional angiography has been superseded by HRCT for diagnoses of anomalous systemic arterial supply. Chest X ray may show a vessle-like opacity. Contrast-enhanced CT scan can be diagnostic. HRCT combined with multiplanar reconstruction, maximum intensity projection and volume reconstruction consistently gain combinations of characteristic findings and perspective [5]. Just like 6 cases reported in present study, orientation and distribution of the anomalous systemic artery were precisely depicted. Most of the time, in anomalous arterial supply without sequestration, one anomalous artery arise from descending thoracic aorta, and feeding the basal segments of the left lower lobe, the venous drainage to the left atrium is via pulmonary veins, and pulmonary artery is absent [2]. Anomalous arterial supply to the right lower lobe other than the left side have been reported [6], so as the left upper lobe [7] and two aberrant arteries [8, 9]. Therefore, although there is usually only a single aberrant artery supply in left, the possibility of another aberrant artery or in other position should be considered.

Treatment is recommended for all patients suffered this anomaly with or without symptoms. Because it has potential risk of hemoptysis and heart failure. Treatment aims to relieve the volume overload on the left ventricle by abolishing the arteriovenous shunting. This can be achieved by lobectomy of the affected lung lobe, ligation of the aberrant artery and therapeutic embolization [10, 11]. Surgical procedures or embolism position would be very carefully selected.

With regard to surgical procedures, left lower lobectomy or basal segmentectomy are still predominant by traditional way or by VATS nowadays in China. The most common complications of surgery are postoperative infection and bleeding. In present six cases, five of them were treated with VATS, only four succeeded and one died of postoperative bleeding.

In reviewing the literature, transarterial embolization has recently been reported more and more in anomalous systemic arterial supply. We found no complication in these reports [12]. Because of less complication and shortened recovery time, the embolotherapy is considered as a minimally invasive, safe, valuable, and more effective treatment compared with surgical intervention [13, 14]. One case reported that a cecum of the stump, which was complication of lobectomy, was repaired by endovascular stent-graft implantation [15]. Although

whether embolization is the first choice for all anomalous systemic supply is still arguable [16], it suggested that embolization was especially suitable for children [17], patients with complicated condition, which including congenital heart disease such as atrial septal defect and scimitar syndrome [18], dual abnormal arteries supply [17], aneurysm [15] and hematologic disease like our case 1.

At last, the limited number of cases in this study were not enough to fully explore the diagnoses and treatment of anomalous systemic arterial supply to basal segments. It is our duty to report and accumulate such cases.

Conclusions

Definite diagnoses can be made by means of high resolution CT, which also plays an important role in the treatment decision. For patients with complicated condition, therapeutic embolization could be a safer alternative.

Disclosure of conflict of interest

None.

Address correspondence to: Benquan Wu, Department of Internal Medicine, Division of Respiratory and Critical Care, The Third Affiliated Hospital of Sun Yat-Sen University, 600 Tianhe Street, Guangzhou 510630, P.R. China. Tel: +86-20-85252226; Fax: +86-20-85252092; E-mail: zswbq@163.com

References

[1] Yamanaka A, Hirai T, Fujimoto T, Hase M, Noguchi M and Konishi F. Anomalous systemic arterial supply to normal basal segments of the left lower lobe. Ann Thorac Surg 1999; 68: 332-338.

[2] Qin J, Wang XL, Bai MJ, Huang SH, Chen XZ and Shan H. Comparison of anomalous systemic artery to left lower lobe and pulmonary sequestration in left lower lobe by computed tomography. Acta Radiol 2015; 56: 1100-1104.

[3] Wu KA, Wu CP and Perng WC. Ethnicity relation to anomalous systemic arterial supply to normal basal segments of the left lower lobe. Clin Imaging 2008; 32: 477-479.

[4] Hubail Z, Eapen R and Lemler M. Anomalous systemic arterial supply to the lungs: an unusual etiology for cardiomegaly. Pediatr Cardiol 2006; 27: 608-611.

[5] Bhalla A, Gupta P, Mukund A, Kumar A and Gupta M. Anomalous systemic artery to a normal lung a rare cause of hemoptysis in adults. Oman Med J 2012; 27: 319-322.

[6] Mautone M and Naidoo P. A case of systemic arterial supply to the right lower lobe of the lung: imaging findings and review of the literature. J Radiol Case Rep 2014; 8: 9-15.

[7] Hwang JH, Kim EY and Park SY. Aberrant bronchial artery to non-sequestrated left upper lobe in massive hemoptysis. Tuberc Respir Dis (Seoul) 2015; 78: 380-384.

[8] Makino T, Hata Y, Otsuka H, Koezuka S, Okubo Y, Isobe K, Tochigi N, Shibuya K, Homma S and Iyoda A. Simultaneous resection of bilateral anomalous systemic supply to the basal segments of the lungs: a case report. J Cardiothorac Surg 2015; 10: 140.

[9] Irodi A, Cherian R, Keshava SN and James P. Dual arterial supply to normal lung: within the sequestration spectrum. Br J Radiol 2010; 83: e86-89.

[10] Canyigit M, Gumus M, Kilic E, Erol B, Cetin H, Hasanoglu HC and Arslan H. Aneurysm of an anomalous systemic artery supplying the normal basal segments of the left lower lobe: endovascular treatment with the amplatzer vascular plug II and coils. Cardiovasc Intervent Radiol 2011; 34 Suppl 2: S126-130.

[11] Mori S, Odaka M, Asano H, Marushima H, Yamashita M, Kamiya N and Morikawa T. Anomalous systemic arterial supply to the basal segments of the lung: feasible thoracoscopic surgery. Ann Thorac Surg 2013; 96: 990-994.

[12] Machida Y, Motono N, Matsui T, Usuda K and Uramoto H. Successful endovascular coil embolization in an elder and asymptomatic case of anomalous systemic arterial supply to the normal basal segment. Int J Surg Case Rep 2017; 34: 103-105.

[13] Gümüştaş S, Akça A, Ciftçi E, Topçu S and Akgül A. A minimal invasive surgical alternative to aberrant systemic arterial supply: coil embolization. Interv Med Appl Sci 2013; 5: 34-38.

[14] Kim JH, Kim SS, Ha KS, Bae J and Park Y. Anomalous arterial supply to normal Basal segment of the right lower lobe: endovascular treatment with the amplatzer vascular plug. Tuberc Respir Dis (Seoul) 2014; 76: 295-298.

[15] Nakagiri T, Sawabata N, Kuratani T and Okumura M. Endovascular stent-graft implantation for a cecum of an aberrant artery from a systemic arterial supply to the basal segment of the left pulmonary lobe. Gen Thorac Cardiovasc Surg 2010; 58: 640-643.

[16] Jackson JE. Regarding "transarterial embolization of anomalous systemic arterial supply to normal basal segments of the lung".

Cardiovasc Intervent Radiol 2017; 40: 634-635.

[17] Jariwala P, Ramesh G and Sarat Chandra K. Congenital anomalous/aberrant systemic artery to pulmonary venous fistula: closure with vascular plugs & coil embolization. Indian Heart J 2014; 66: 95-103.

[18] Yamakawa H, Shimizu K, Michimoto K, Kameoka Y, Kang R, Yoshida J, Yamada M, Yoshida M, Ishikawa T, Takagi M and Kuwano K. Transcatheter embolization for hemoptysis associated with anomalous systemic artery in a patient with scimitar syndrome. Springerplus 2015; 4: 422.